#### **IMMEDIATELY NOTIFIABLE TO DOH: 1-877-539-4344**

Potential Bioterrorism Agent: Category A

# **Botulism**

# (Foodborne, Wound, Infant, Other)

# 1. DISEASE REPORTING

# A. Purposes of Reporting and Surveillance

- 1. To assist in the diagnosis of potential cases and facilitate prompt administration of either antitoxin or botulism immune globulin when indicated.
- 2. For foodborne botulism, to identify contaminated food(s) and to prevent further exposures.
- For foodborne botulism, to identify and assure the proper evaluation and care of other persons who may be at immediate risk of illness because they have already eaten the implicated food.
- 4. For wound botulism, to alert others at risk regarding the importance of promptly identifying illness and obtaining medical care.

### **B.** Legal Reporting Requirements

- 1. Health care providers: immediately notifiable to local health jurisdiction.
- 2. Hospitals: immediately notifiable to local health jurisdiction.
- 3. Laboratories: **immediately notifiable to local health jurisdiction**. Specimen submission is required.
- 4. Local health jurisdictions: **suspected and confirmed cases are immediately notifiable** to the Washington State Department of Health (DOH) Communicable Disease Epidemiology Section (CDES) (1-877-539-4344).

#### C. Local Health Jurisdiction Investigation Responsibilities

- 1. Call CDES immediately to report suspect cases and discuss the need for antitoxin or botulism immune globulin intravenous (human) (BIG-IV).
- 2. Determine the most likely source of the exposure and prevent others from being exposed.
- 3. Facilitate the transport of appropriate specimens to the Washington State Department of Health Public Health Laboratories (PHL).
- 4. Report all *probable* and *confirmed* cases of foodborne botulism to CDES through the Public Health Issues and Management System (PHIMS) using the **foodborne botulism** case report form (www.doh.wa.gov/notify/forms/bot\_food.doc).
- 5. Report all *confirmed* cases of wound botulism to CDES through PHIMS using the **wound** botulism case report form (www.doh.wa.gov/notify/forms/bot\_wound.doc).
- 6. Report all *confirmed* cases of infant botulism to CDES through PHIMS using the **infant botulism** case report form (<u>www.doh.wa.gov/notify/forms/bot\_infant.doc</u>).

Last Revised: August 2008 Page 1 of 11 7. Report all other *confirmed* cases of botulism through PHIMS as Botulism, Other including adult colonization botulism, inhalational botulism and botulism from an unknown source.

# 2. THE DISEASE AND ITS EPIDEMIOLOGY

# **Background**

Botulism is a neurological disease caused by botulinum toxin. Although all types described below are potentially fatal and demand emergency medical intervention, only foodborne and inhalational botulism are public-health emergencies.

- <u>Foodborne botulism</u> occurs when a person ingests toxin, which leads to illness within a few hours to days. Outbreaks of foodborne botulism have potential to be a public health emergency because the contaminated food may be eaten by other people.
- <u>Wound botulism</u> is a rare disease that occurs when wounds infected with *C. botulinum* secrete the toxin.
- <u>Infant botulism</u> occurs each year in a small number of susceptible infants who harbor *C. botulinum* in their intestinal tract. It occurs when an infant ingests spores of *C. botulinum*, which in turn colonize the intestinal tract and produce toxin.
- <u>Adult colonization botulism</u> is an even rarer type of intestinal colonization reported only a few times in the literature. It involves intestinal colonization with *C. botulinum* in a person older than one year of age. In the small number of these cases, most patients had a history of gastrointestinal surgery or illness, such as inflammatory bowel disease, which might have predisposed them to enteric colonization. No other specific risk factors have been identified.
- <u>Inhalational botulism</u> occurs after inhalation of botulism toxin. This does not occur naturally. There have been only three reported cases in humans world-wide, associated with laboratory exposure.

# A. Etiologic Agent

Botulism is caused by a toxin produced by the gram-positive bacillus *Clostridium botulinum*. The toxin binds irreversibly at the neuromuscular junction. *C. botulinum* forms spores which can survive under a wide range of adverse environmental conditions including boiling. The higher temperatures (>120.5°C/250.5°F) that can be achieved under pressure (e.g., in an autoclave or *properly functioning* home pressure cooker) are sufficient to kill even spores.

Spore germination and bacterial growth occur only under anaerobic and low-acid to non-acidic (generally pH>4) conditions. Toxin is produced as the bacteria multiply. Botulinum toxin is heat-labile and can be inactivated by boiling for ten minutes. There are seven types of botulinum toxin, designated A–G. Types A, B, and E are the most common sources of human disease; type E is highly associated with marine products (fish, seafood, or marine mammal meat). F is very rare, and C, D and G are not known to cause human illness. The toxin is a potential agent of bioterrorism.

# **B.** Description of Illness

Symptoms of botulism may include the "4 D's" – dysphagia (difficulty swallowing), dry mouth, diplopia (double vision), and dysarthria (difficulty articulating words) as well as blurred vision, ptosis (drooping eyelids), and muscle weakness. Symptoms reflect a descending, symmetrical flaccid paralysis that starts with the facial muscles and may progress downward. Respiratory distress may ensue if the muscles of breathing are compromised. Mental alertness and peripheral sensation are typically maintained. Neurologic symptoms may be preceded or accompanied by mild gastrointestinal disturbance such as constipation, vomiting, or diarrhea. The severity of symptoms and the rate of progression are highly variable, depending on dose and other factors. In severe cases, patients may survive only after months on a ventilator.

Botulism is frequently misdiagnosed, most often as polyradiculoneuropathy (Guillain-Barré or Miller-Fisher syndrome), myasthenia gravis, or other diseases of the central nervous system.

In infants with intestinal botulism the first sign is often constipation, followed by lethargy, listlessness, a weak cry, ptosis, difficulty feeding (weak or absent sucking response) leading to weight loss, and generalized weakness (the "floppy baby" syndrome). The infant may present with "failure to thrive."

# C. Botulism in Washington State

During the last 10 years, DOH has received 0–2 reports of foodborne botulism per year, 0–6 reports of intestinal botulism per year and 0–7 reports of wound botulism per year.

Recent foodborne botulism cases in Washington have been associated with improperly home-canned vegetables. Wound botulism is most frequently associated with injection drug use, particularly black tar heroin.

#### D. Reservoirs

C. botulinum spores are common in soil and elsewhere in the environment including on vegetables.

#### E. Modes of Transmission

Modes of transmission for foodborne, wound and infant botulism are described here.

#### 1. Foodborne botulism

Foodborne botulism is caused by ingestion of preformed toxin. Typically, implicated foods have been low acid, home-canned foods that have not been processed adequately during canning and not heated before consumption. Rarely, commercial products are implicated, usually after some breakdown in standard canning procedures has occurred. Examples of implicated foods include:

- home-canned asparagus, beans, and other vegetables (including low-acid tomatoes), usually canned by the water-bath method;
- fish that has been improperly canned, dried, or stored;
- sausage or other prepared meats that are improperly processed (inadequate sodium nitrite) and improperly stored;

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- chopped garlic or eggplant bottled in oil;
- among Alaska Natives, traditionally preserved foods including fermented (putrefied) whale blubber, salmon heads, salmon eggs, and other delicacies
- in 2007, commercially canned chili.

# 2. Wound botulism

Wound botulism results from a local C. *botulinum* infection in devitalized tissue at a wound site, where semi-anaerobic conditions occur. As with intestinal botulism, the toxin is produced *in situ* and disseminated in the blood. Wound botulism is increasingly reported, especially due to subcutaneous injection ("skin popping") of black-tar heroin.

#### 3. Infant botulism

Intestinal botulism occurs when *C. botulinum* spores, ingested in food or soil, germinate in an intestine that does not have a mature flora. Botulinum toxin is then produced in the intestine and enters the bloodstream. Although in the past an association was suggested for intestinal botulism in infants and honey, recent cases have rarely consumed honey.

Most cases occur in infants less than three months old (almost always under six months old). Cases occur in both breast fed and formula fed infants.

#### F. Incubation Period

- 1. Foodborne botulism: The incubation period for foodborne botulism varies from 12 hours to several days, but is usually 12–36 hours. A short incubation is associated with more severe disease.
- **2.** Wound botulism: The incubation period can be up to two weeks or longer.
- **3. Infant botulism:** The incubation period is unknown.

#### G. Period of Communicability

Botulism is not communicable from person to person.

#### H. Treatment

Treatment should never be delayed pending laboratory confirmation of the diagnosis. All patients require close monitoring of ventilatory status, and aggressive supportive therapy is required in severe cases. Some patients have recovered completely only after months on a ventilator. Additional therapies depend on the type of botulism and are outlined below.

#### 1. Foodborne botulism

Foodborne botulism is treated with botulism antitoxin. Although the antitoxin cannot reverse symptoms caused by irreversibly bound toxin, treatment will halt the progression by removing free toxin if given promptly after exposure. Antitoxin therapy should never be delayed pending laboratory confirmation of the diagnosis, which often takes several days. The divalent (anti-A and -B) and trivalent (anti-A, -B, and -E) antitoxins are purified from horse serum, and about 9% of recipients suffer allergic reactions. A skin test using the vial is always indicated before administration of antitoxin; the directions are in the antitoxin package insert. If no hypersensitivity reaction to the skin test occurs,

the full dose (1 vial) may be given intravenously. Trivalent antitoxin is usually used for foodborne botulism unless marine products can definitely be ruled out as the source of exposure.

CDC controls the distribution of botulinum antitoxin, which is stocked at United States Public Health Service Quarantine Stations throughout the country. A Quarantine Station is located at SeaTac Airport in Seattle. If antitoxin use is being considered,

<u>IMMEDIATELY</u> consult with CDES (1-877-539-4344). Personnel at CDC and SeaTac Quarantine station can arrange to have the antitoxin transported to the hospital where the patient is being treated.

# 2. Wound botulism

Wound botulism is treated with botulism antitoxin. Antitoxin should be administered as for foodborne botulism, typically as the divalent form. Debridement of the wound is indicated to remove devascularized tissue that provides the anaerobic conditions required for growth of *C. botulinum*. There is a theoretical reason to postpone debridement until after antitoxin administration to avoid further toxin release. If antitoxin use is being considered, **IMMEDIATELY** consult with the CDES (1-877-539-4344) as for food botulism. Antimicrobial therapy should also be considered.

#### 3. Infant botulism

Infants may do well with supportive care or may need weeks of hospitalization and mechanical ventilation. A human-derived botulism hyper-immune globulin (BIG-IV) was approved by FDA in 2003 for treatment of infants. Though the cost is substantial, its use may be cost-effective. A randomized, double-blind, placebo-controlled trial of BIG-IV found a 3-week reduction in the mean length of hospital stay with an accompanying reduction in the mean hospital charges. Consultation or BIG-IV can be obtained from the California Department of Health Services by **IMMEDIATELY** calling the 24-hour number at 510-231-7600. Also contact CDES to arrange for testing.

#### 4. Adult colonization botulism

Horse-derived antitoxin is used to treat adult colonization botulism. More than one dose of antitoxin may be required. If antitoxin use is being considered, **IMMEDIATELY** consult with CDES (1-877-539-4344) as for food botulism.

### 3. CASE DEFINITIONS

# A. Case Definition for Foodborne Botulism

- 1. Clinical Criteria for Diagnosis: Ingestion of botulinum toxin results in an illness of variable severity. Common symptoms are double or blurred vision, and difficulty swallowing or speaking. Descending symmetric paralysis may progress rapidly.
- 2. Laboratory Criteria for Diagnosis: Detection of botulinum toxin in serum, stool, or patient's food, or isolation of *Clostridium botulinum* from stool.

#### 3. Case Definition

a. Probable: A clinically compatible case with an epidemiologic link (e.g., ingestion of a home-canned food within the previous 48 hours).

Last Revised: August 2008 Page 5 of 11 b. Confirmed: A clinically compatible case that is laboratory confirmed or that occurs among persons who ate the same food as persons who have laboratory-confirmed botulism.

#### **B.** Case Definition for Wound Botulism

- 1. Clinical Criteria for Diagnosis: An illness resulting from toxin produced by *Clostridium botulinum* that has infected a wound. Common symptoms are double or blurred vision, and difficulty swallowing or speaking. Descending symmetric paralysis may progress rapidly.
- 2. Laboratory Criteria for Diagnosis: Detection of botulinum toxin in serum or isolation of *C. botulinum* from wound.

#### 3. Case Definition

a. Confirmed: a clinically compatible case that is laboratory confirmed in a patient who has no suspected exposure to contaminated food and who has a history of a fresh, contaminated wound during the 2 weeks before onset of symptoms.

# C. Case Definition for Infant Botulism

- 1. Clinical Criteria for Diagnosis: An illness of infants, characterized by constipation, poor feeding, and "failure to thrive" that may be followed by progressive weakness, impaired respiration, and death.
- 2. Laboratory Criteria for Diagnosis: Detection of botulinum toxin in serum or stool, or isolation of *C. botulinum* from stool.
- 3. Case Definition
  - b. Confirmed: a clinically compatible case that is laboratory-confirmed, occurring in a child aged < 1 year.

#### D. Case Definition for Botulism, Other

- 1. Clinical description: See Foodborne Botulism.
- 2. Laboratory criteria for diagnosis: Detection of botulinum toxin in clinical specimen or isolation of *Clostridium botulinum* from clinical specimen.
- 3. Case classification
  - a. *Confirmed:* a clinically compatible case that is laboratory confirmed in a patient aged greater than or equal to 1 year who has no history of ingestion of suspect food and has no wounds.

# 4. DIAGNOSIS AND LABORATORY SERVICES

# A. Diagnosis

Presumptive tests for botulism toxin may be completed in one day but the confirmatory assay used to detect botulism toxin and cultures for *C. botulinum* take several days to complete. Therefore, treatment should never be delayed pending laboratory confirmation of the diagnosis.

1. **Foodborne botulism:** Diagnosis is made by detecting botulism toxin in serum, stool, or implicated food or by culturing *C. botulinum* from stool. Vomitus or gastric aspirate can be tested for toxin if obtained within a few hours of food ingestion.

- **2. Wound botulism:** Diagnosis is made by detecting botulism toxin in serum or by culturing *C. botulinum* from the infected wound. Stool should be obtained in addition to rule out foodborne botulism.
- **3. Infant botulism:** Diagnosis is made by detecting botulism toxin or by culturing *C. botulinum* from stool. In contrast to foodborne and wound botulism, the toxin is rarely detected in serum.

#### B. Tests Available at PHL

DOH Public Health Laboratories perform presumptive (ELISA) and confirmatory botulism toxin assays, and cultures clinical or environmental specimens (e.g., food) for *C. botulinum*. Consult with CDES to arrange for testing.

# C. Specimen Collection

Collect serum prior to the administration of antitoxin. Once antitoxin is administered, it binds to free toxin in the blood and prevents it from being detected by assay. Obtain all other clinical specimens early in the course of illness and if possible before administration of antitoxin. Collect all clinical specimens in sterile leak-proof containers.

- 1. For <u>stool</u> testing, submit at least 15 grams of stool, if possible 50 grams (about ping-pong ball sized). If the patient is constipated, which is common with botulism, a small amount of sterile, nonbacteriostatic fluid may be used for an enema. For post-mortem testing, collect multiple 15 gram specimens from different parts of the small and large intestine.
- 2. For <u>serum</u> testing, submit at least 5 ml of serum (not blood), if possible 10–15 ml for an adult and 3–4 ml for an infant. Serum left over from other testing may be used.
- 3. For <u>food</u> testing, send as much implicated food as possible from the source suspected of being consumed. Typically only opened jars or containers are tested. Rinsed canning jars have tested positive in the past. Each item should be packed individually in sterile unbreakable containers with secure seals.
- 4. For wound testing, send swabs for culture in anaerobic transport medium.
- 5. For gastric aspirate or vomitus, send at least 20 ml.

All specimens should be kept refrigerated (*not frozen*) during storage and transport. Use cold packs to maintain a shipping temperature of 4°C (39°F). Specimens must be properly packaged using guidelines for shipping and packaging of diagnostic specimens. Be sure to use absorbent material around the primary container, particularly food specimens, which could have high levels of toxin and present a danger if there is leakage. Include a completed DOH Reference Bacteriology form 302-006a (available at: <a href="http://www.doh.wa.gov/EHSPHL/PHL/Forms/ReferenceBacteriology.pdf">http://www.doh.wa.gov/EHSPHL/PHL/Forms/ReferenceBacteriology.pdf</a>) with all specimens.

# 5. ROUTINE CASE INVESTIGATION

If you identify more than one case of botulism without an obvious source of infection, consider the possibility of an outbreak associated with a commercial product or an act of bioterrorism and call CDES <u>IMMEDIATELY</u> at (877-539-4344) (see Section 7).

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# A. Evaluate the Diagnosis and Arrange for Treatment

Obtain information from the provider and others regarding the patient's history and physical exam findings, particularly neurologic exam findings (e.g., cranial nerve function) and call CDES <u>immediately</u> (877-539-4344) to discuss the case. For suspected foodborne or wound botulism, an epidemiologist at CDES will call CDC to release botulism antitoxin if needed. For suspected intestinal botulism in an infant, the provider should be immediately referred to the California Department of Health Services (24-hour number 510-231-7600). Treatment should never be delayed pending laboratory confirmation of the diagnosis since laboratory confirmation can take several days and treatment needs to be given immediately.

While the antitoxin or BIG-IV is being released, arrange for diagnostic specimens to be sent to PHL (see Section 4 above).

### **B. Identify Source of Infection**

- 1. <u>Foodborne Botulism</u>: Interview the case and others who may be able to provide pertinent information about foods eaten. A home visit is strongly recommended when homecanned foods are implicated, or if the source is not readily apparent. Identify the following products that were consumed in the week prior to onset of symptoms:
  - a. Home-canned, vacuum packed, or traditionally preserved foods. The most suspect foods are those that were eaten less than two days before onset, those that are low in acid (vegetables, fish, and meat), and those that were not eaten by other persons who remain well. (Keep in mind, however, that some cases may develop symptoms several days after the index case.) Identify and collect all remaining jars of the homecanned foods.
  - b. Commercially canned or vacuum packed foods or mishandled commercial products (e.g., refrigeration not maintained); such products are implicated only rarely. For implicated foods, determine the brand, manufacturer, package size, lot number, and place and date of purchase. When a commercial product or environmental exposure is implicated, see Section 6. Controlling Further Spread
  - c. Sausage, preserved or traditionally preserved meats, and inadequately refrigerated meats; such products are implicated only rarely.
  - d. Preserved or traditionally prepared fish and marine products.
  - e. Items stored in oil (e.g. onions, garlic) or foil (e.g. baked potatoes.)
  - f. With the identification of more than one case of botulism without a likely source of exposure consider the possibility of intentional exposure through deliberately contaminated food and interview the case and others who may be able to provide pertinent information about possible exposures such as locations where food and water were consumed, particularly at gatherings and public events.
- 2. Wound Botulism: Ask the patient about illicit drug use. Specifically, ask about the type of drugs used and how the drugs are used (e.g., injected into veins, injected into tissues, snorted, etc.). It is often difficult to specifically identify sources of heroin. Testing of heroin or drug paraphernalia is not encouraged. In addition to illicit drug use, interview regarding potential foodborne exposures.

- 3. <u>Infant Botulism</u>: No specific exposures are well described. Although honey was associated with intestinal botulism in the past, it is rarely implicated in cases.
- 4. <u>Botulism</u>, <u>other</u>: If the type of botulism cannot be determined, identify public events and gatherings that the case attended.

# C. Identify Potentially Exposed Persons

- 1. Obtain the name, address, and telephone number of every person who may have eaten the suspected food item or shared an environmental exposure.
- 2. Obtain the organization name, contact telephone number, and attendance lists (particularly e-mail or telephone lists) for every suspected gathering, public event, or other shared environmental exposure.
- 3. Obtain the name, address, and telephone number of every person who may have the suspect home-processed food in his or her possession.

#### **D.** Environmental Evaluation

If a restaurant is implicated, conduct an immediate inspection to identify home canned or mishandled product in the facility. If an environmental exposure is suspected, contact CDES for assistance with sampling.

# 6. CONTROLLING FURTHER SPREAD

#### **A. Infection Control Recommendations**

Hospitalized patients should be treated with standard precautions.

# **B.** Case Management

No case follow-up is needed after the case receives treatment.

# C. Contact Management

No contact follow-up is needed since botulism is not transmitted from person to person.

# D. Management of Other Exposed Persons

- 1. <u>Foodborne botulism</u>: If reachable within six hours of exposure, other persons who have eaten implicated food should be purged and given gastric lavage to remove any unabsorbed toxin. They should be monitored for signs of botulism at least twice daily for three days, and instructed to seek medical care immediately should symptoms develop.
- 2. <u>Wound botulism</u>: When possible, provide education to risk groups and to health care providers serving them regarding typical symptoms of botulism and the importance of rapid diagnosis and treatment. Potential routes for education include needle exchange programs and urban hospital emergency departments.

#### E. Environmental Measures

- 1. Home-Canned Food Implicated
  - a. Samples of any implicated home-canned food should be sent to the PHL for testing, and the remainder destroyed. To avoid endangering trash haulers or others, these foods should be autoclaved before discarding; as an alternative, bring contents of

containers to a full boil for a least ten minutes. Any empty containers should be likewise boiled.

- b. The person who prepared the home-canned food should be thoroughly instructed in proper canning techniques.
- 2. Commercial Products Implicated: When a commercial product is implicated as the source of intoxication, CDES should be notified **immediately.** They will coordinate follow-up with relevant outside agencies (FDA, USDA, CDC, etc.)

# 7. MANAGING SPECIAL SITUATIONS

#### A. Outbreak

With the identification of more than one case of botulism without an obvious source of exposure, consider the possibility of a contaminated commercial food product. **In such situations call CDES IMMEDIATELY: 1-877-539-4344.** The cases will need to be extensively interviewed to identify possible exposures such as locations where food and water were consumed, particularly at gatherings and public events.

#### **B.** Bioterrorism

C. botulinum toxin has been classified as a possible agent of bioterrorism because it is extremely potent and lethal. The toxin is also easy to produce and transport, and affected individuals often need extensive and prolonged intensive care. Aerosol dissemination through air or on food would be the most likely mode of spread. Aerosol dissemination could produce many cases in a geographic area. Therefore, inhalational botulism produced by an act of bioterrorism should be considered for two or more botulism cases which are linked temporally and geographically but without a likely foodborne or drug exposure. In such situations call CDES IMMEDIATELY: 1-877-539-4344. The cases will need to be extensively interviewed to identify possible exposures such as gatherings, public events, specific geographic locations, large buildings or shopping areas, and public transportation.

# 8. ROUTINE PREVENTION

#### A. Vaccine Recommendations

None

#### **B. Prevention Recommendations**

- 1. Foodborne botulism
  - Persons who do home canning should follow strict hygienic procedures to reduce contamination of foods.
  - Oils infused with garlic or similar moist flavoring should be refrigerated.
  - Potatoes which have been baked while wrapped in aluminum foil should be kept hot until served or refrigerated.
  - Because the botulism toxin is destroyed by high temperatures, persons who eat risky home-canned foods (i.e., low acidic, non-pickled foods) should consider boiling the

Last Revised: August 2008 Page 10 of 11 food for ten minutes before eating it to ensure safety. Pickling, sugar syrup, or sufficient brining should prevent the growth of *C. botulinum*.

• Instructions on safe home canning can be obtained from county extension services or from the United States Department of Agriculture.

#### 2. Wound botulism

- Wound botulism can be prevented by promptly seeking medical care for infected wounds and by not using injectable street drugs.
- IV drug users and health care providers serving them should be educated regarding typical symptoms of botulism and the importance of rapid diagnosis and treatment. Potential routes for education include needle exchange programs and urban hospital emergency departments.

#### 3. Infant botulism

- Because honey can contain spores of *Clostridium botulinum* and this has been a source of infection for infants, children less than 12 months old should not be fed honey raw or otherwise.
- Honey is safe for persons one year of age and older.

# **ACKNOWLEDGEMENTS**

This document is a revision of the Washington State Guidelines for Notifiable Condition Reporting and Surveillance published in 2002 which were originally based on the Control of Communicable Diseases Manual (CCDM), 17<sup>th</sup> Edition; James Chin, Ed. APHA 2000. We would like to acknowledge the Oregon Department of Human Services for developing the format and select content of this document.

# **UPDATES**

Section 2F: The incubation period for wound botulism can be up to two weeks or longer.